CHOREA:

A STUDY IN CLINICAL PATHOLOGY.

BY

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PRECISELY as at one time paralysis was supposed to be a disease, so at a later period was chorea believed to be a distinct affection. Indeed, many of the profession still use the term as though it were the name of a distinct ailment, and most of the treatises upon nervous diseases have a chapter upon chorea as a separate affection. It is, however, perfectly clear that the word should be employed as analogous to, or having equal rank with, paralysis or tremor. The term choreic movements is unnecessary; or, if it still be used, the term chorea should be considered to represent that condition in which choreic movements occur.

Of the various diseases or conditions in which choreic movements occur, I shall first speak of that which is known as *post-paralytic chorea*, or sometimes *post-hemiplegic chorea*, and begin by a brief record of a case now under my care.

Miss W., aged 29. At nine months of age, probably after improper feeding, she was seized with a violent convulsion. These convulsions recurred at brief intervals until she was about five years old, when she had an exceedingly bad one, lasting all night. During this time complete right-sided hemiplegia appeared. Three or four months afterwards she began to have movements in the right arm. These have continued ever since. She had convulsions after this frequently, until about the age of fifteen, when, menstruation setting in, the convulsions became more infrequent, and she has not had one since 1875.

Present condition, a well-nourished, healthy-looking woman. Right side of the face somewhat smaller than the left side of the face, and the left angle of mouth, on smiling, going up very distinctly more than the right. Every time she winks there is a very curious local spasm, covering about the size of half a dollar, in the right side of the chin. Occasionally a slight tremulousness about the right corner of the mouth. The spasms are chiefly in the arm, causing the hand to vibrate rapidly. There are also spasms of the forearm, causing disorderly movements of the fingers; right hand registered on the dynamometer 45, left hand 75; while the right-arm muscles are apparently bigger than the left. There is a perpetual perspiration on the right arm. The deltoid muscle evidently takes part in the spasm, and is exceedingly large. There is slight loss of sensation in the right arm. By fixing the hand against the body, immediately under the right breast, she is able to do such work as crochetting. Complains much of pain in the upper arm, especially in the region of the deltoid. This pain is worse on getting up in the morning. She does not know about spasms during sleep, but generally lies on her arm. General intelligence and health perfect.

This case is especially interesting on account of the co-association of winking with a very peculiar local spasm involving only a few fibres of the facial nerve. In some way the oculo-motor centres have become linked with a few cells of one facial nucleus, so that one reflex nerve-discharge influences the three centres, a combination very difficult to explain.

This case illustrates many of the peculiarities of this so-called post-hemiplegic chorea, but it seems to me as absurd to speak of this condition as a distinct affection as it would be of contractures occurring after an apoplexy. There are, however, certain very curious facts connected with the affection worthy of here being noted. Very generally there is associated with it a loss of sensibility; then again the movements are almost always confined to the upper extremities. Although

in some of the reported cases there was a lack of control over the movements of the leg, I cannot recall a single instance in which there were in the leg decided spasmodic movements such as occur in the arm of Miss W.

The following case of chorea now under my care at the University Hospital is in many respects closely allied to the cerebral chorea of early childhood, and seems worthy of being noted, although the history of it is very imperfect. The patient is a young man 19 years of age, well nourished, muscular, who for the last three years has followed the occupation of driving trotting-horses, both in training and races. He states that his present difficulty began when he was four years of age, and that two years since the movements were greater than they are at present: except for the movements, the health is perfect, and the muscular power of the whole body is much above the average. The movements are confined to the upper extremities, including the group of shoulder muscles; they are worse in the right than in the left arm; are constant during the day; do not interfere with his going to sleep, and always cease at such time. They are greatly increased by excitement, or when he attempts any movement with his arms under observation. They are much lessened by any steady muscular act, such as driving a hard-mouthed horse or firmly grasping a chair. They are mostly typical choreiform movements, but at times when he is very much excited become so marked as to appear almost like voluntary movements. The patella reflexes are very active, and there is in the right arm a slight but distinct finger-reflex when the tendons on the wrist are tapped.

The following extract from my note-book shows that the loss of sensibility is very slight, and I am not sure whether the lesion is spinal or cerebral, although inclined to the latter view. "Lowest point of separation of points. *Right arm*—back and front of forearm, 7.5 cm.; palm, 0.6 cm.; tips of fingers, 0.4 cm.; back of fingers, 0.5 cm.; region of biceps and of deltoid, 7 cm. *Left arm*—upper and forearm as on other side; tips of fingers, 0.2 cm.; back of fingers, 0.5 cm."

There are two classes of these cases, one in which the choreic movements develop at the same time as the palsy, or even before it, and one in which they first appear long after the loss of power. It is very desirable that close microscopical examinations should be made in the latter class of cases, as it is very possible that the symptoms may be connected with secondary degenerations. Such connection is established for post-paralytic contractures, and there is reported in the *Hospital Gazette* (vol. iv. p. 366) a case in which such contractures co-existed with choreic movements, and in which there was almost certainly secondary degeneration of the nerve-centres.

The seat of the lesion in post-hemiplegic chorea, as first stated by Charcot, and especially developed in the thesis of his pupil, Raymond, is in the posterior part of the internal capsule, in the immediate neighborhood of the lenticular nucleus and optic thalamus. The immediate band of fibres of the corona radiata especially involved is in front of that connected with and covering the posterior end of the optic thalamus. This region, it will be remembered, is a very distinct one, having its own artery, the posterior optic. Although in many cases of post-hemiplegic chorea the lesion is located in the spot designated by the great French neurologist, such location is not invariable. M. Demange (*Rev. de Méd.*, March, 1883, p. 377), after reporting a case with Charcot's lesion, records one in which there was violent post-hemiplegic chorea, and in which the lesion was situated in the convolutions. It is a very interesting feature in this case that the choreic movements were preceded by epileptiform crises, which ceased when the choreic movements developed. The choreic movements themselves also disappeared before death. In two cases of cerebral syphilis, with presumably cortical lesion, I have seen a violent choreiform spasm of the face replace epileptiform convulsions, and there is a close analogy between post-hemiplegic chorea and Jacksonian epilepsy. Dr. Demange reports a case in which hemiplegia was associated with severe tremblings, like those of paralysis agitans, and the lesion was situated in the lenticular nucleus. This form of tremor may, however, be considered distinct from true post-hemiplegic chorea, but in the *Bulletin of the Anatomical Society of Paris*, 1879 (vol. liv. p. 748), is recorded a case in which a true post-paralytic chorea was found to depend upon a softening of the brain on the level of the first convolution, involving the whole thickness of the external capsule, as the sole lesion. This case separates itself from those of Charcot in the complete preservation of sensibility, and as the choreic movements developed months after the coming on of the paralysis, it is fair to suspect the presence of an unobserved secondary degeneration.

A very important fact in the natural history

of post-hemiplegic chorea is that it may exist in its most typical form without organic lesion, as is shown by the case reported in the *Bulletin of the Anatomical Society* (vol. lv. p. 150). A woman, aged 49, with a past hysterical history, and also one of chronic iritis and arterial atheroma, but who denied syphilis, suffered paresthesias in the right arm and leg, with vertigo, lasting for some days. She then had a sudden right hemiplegia, without loss of consciousness. The paralysis was at first complete, but eight days later she began to walk a little. Two months after this she had a second attack. Several months subsequent to this she entered the hospital. At that time she had complete right-sided hemiplegia both of common sensibility and of the special senses, and a partial right motor hemiplegia, with very marked choreic movement of the right arm. Neither the right leg nor the left side were in movement. She died of an intercurrent pneumonia, and after death a very careful microscopic examination of the brain failed to detect any lesion. During life a murmur of an aortic insufficiency had been heard, and after death the appropriate lesion was found. It seems to me that the reporter of this history is correct in affirming that during life it was impossible to determine that the case was not one of cerebral disease. This case brings up clearly the possibility that in some even of these prolonged post-paralytic choreas there may be no organic affection.

Not only may post-hemiplegic, but also other forms of chorea be imitated by hysteria. I have not rarely seen cases offering the ordinary manifestations of Saint Vitus's dance of childhood in extremely hysterical young females, and believe them to be of pure hysterical origin.

The so-called *electric chorea* of French writers is evidently, in many cases, hysterical. It consists of very rapid, rhythmical movements of the head or other portions of the body, occurring at intervals, as though in a series of shocks. A very interesting case is reported in the Thesis of Andrè Guertin, Paris, 1881, which shows that this electric chorea may coexist with general choreic movements, exactly resembling those of the Saint Vitus's dance. In some cases of hysteria the movements are entirely rhythmical, and it is perfectly evident all forms of choreic spasms may be produced by hysteria.

The truth of the following propositions may be considered established by facts which have been already adduced :

First.—Choreic movements may be the result of organic brain-disease.

Second.—Choreic movements exactly simulating those of organic brain-disease may occur without any appreciable disease of the nerve-centres.

Third.—General choreic movements, as well as the bizarre forms of electric and rhythmical chorea, may occur without any organic disease of the nervous system.

To these propositions may be added a *fourth*, viz.: Choreic movements may be the result of a peripheral irritation, or, in other words, may be reflex.

As proof of the last proposition, I would cite the following cases :

Dr. C. Fischer reports (*Zeitschrift f. Wundärzte*, 1853, Bd. 6, p. 89) the case of a young peasant girl, who suffered from a violent periodontitis, and a futile attempt at a removal of the tooth. The operation was followed by the formation of an abscess, and marked unilateral chorea, which lasted two years, when Dr. Fischer removed the roots of the tooth, and the movements ceased at once.

Dr. R. Fischer (*Oester. Med. Wochenschrift*, 1841, p. 46) reports a case in which a general chorea ceased at once upon the expulsion of a tapeworm. Dr. Edmond Censier records a case similar to this in the *Gaz. Méd.-Chir. de Toulouse*, 1877, p. 43. The chorea was so violent as to threaten life, and had persisted several months, notwithstanding treatment. Amelioration began five days after the expulsion of the parasite, was very rapid, and resulted in complete cure.

In the *Journ. de Méd. et de Chirurg.*, Paris, 1841, is the record of a case in which chorea of one month's duration was immediately arrested by the expulsion of lumbricoid worms.

M. Borelli reports (*Bulletin de la Soc. de Chirurgie*, 1852, p. 292) the case of a boy thirteen years old, in whom a violent chorea, which had resisted all treatment for six months, was cured by the removal of a neuromatous tumor from beneath the foot. The movements became less the day after the operation, and by the fourth day had ceased entirely.

Further citations of cases might readily be made, and especially an abundance of opinion be obtained from recognized authorities, as showing the occasional dependence of chorea upon the presence of intestinal worms ; but I think enough has been here said for the purpose. The chorea which occurs during pregnancy is also probably of reflex origin.

There is a form of chorea which is not un-

common in the young of certain of our carnivorous domestic animals. I have never seen a case in herbivora; but Prof. Huidekoper, of the Veterinary Department of the University of Pennsylvania, informs me that he has treated the disease in calves.

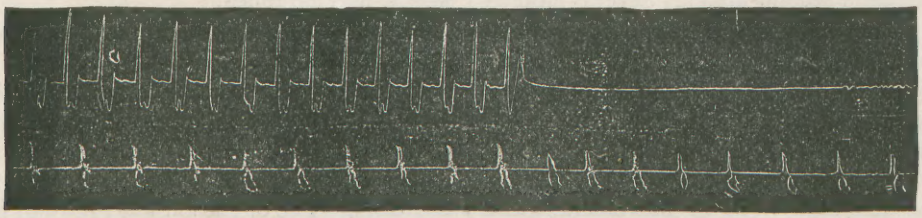
The relations of this disease with the chorea of childhood have been considerably discussed, with difference of conclusions. I shall postpone the consideration of this question until after a study of the animal chorea, of which I have seen a number of examples. It is impossible to experiment, except within very narrow limits, upon human beings, and hence it is only by the light of investigations upon the lower animals that we can hope to come to any very definite conclusions. In such investigations, the points to be inquired into are as to the seat and nature of the lesion, and as to the effect of external irritations upon the symptoms themselves.

The first study of this affection directed towards the discovery of the seat of the lesion of which I have knowledge is that of Prof. Chauvet (*Mem. et Compt. Rend. Soc. de Sci. Méd. de Lyon*). I have never seen this paper, but it is quoted as affirming that the movements persist in the hind legs after section of the spinal cord.

In April, 1869, M. Carviler presented to the Société de Biologie (*Compt. Rend. Soc. Biolog.*, 1869, p. 154) an account of some experiments upon a choreic dog, with persistence of movements after section of the cord in the middle dorsal region. The peculiarity in this report is the assertion that after section of the cord the movements of the face, front and hind legs were perfectly isochronous. It seems to me that it is scarcely possible for such isochronous movements to occur when the spinal cord has been completely severed, as the simultaneousness of movement could only depend upon nerve-waves running down an uninterrupted cord. It is therefore a natural suspicion that the cord was not entirely divided.

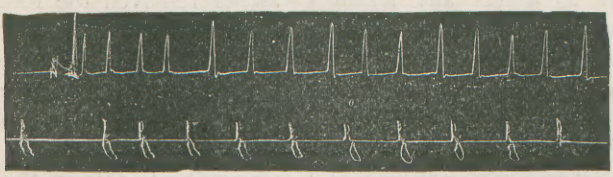
The most elaborate study of the subject is that of Legros and Onimus (*Journ. de l'Anat.*

A



Tracing 1.—A, the point where the drum was stopped and allowed to again revolve when etherization was complete.

A



Tracing 2.—A, the point where drum was started so soon as effect of ether had passed off.

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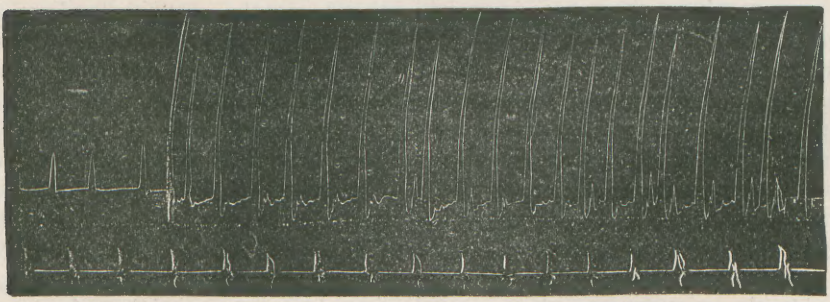


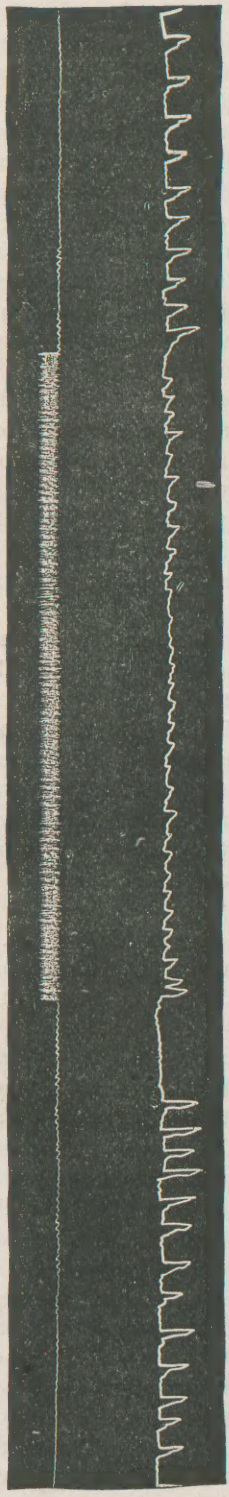
Tracing 3.—A, the point where drum was stopped until the effect of shock had entirely passed by, showing that the movements of the feet were much greater after than before the section. Compare with Tracing 1.

et de la Physiolog., 1870–71, p. 403). These observers found that the movements are not synchronous with the beat of the heart, and do not vary *pari passu* with the cardiac pulsation; that they are not arrested by section of the spinal cord, but are to a certain extent by the action of the cerebrum, for a sudden loud noise would for a time cause them to cease; that they are very sensibly affected by chloral; that division of the sensitive roots of an isolated section of the cord lessens, but does not arrest, the movements; and that a galvanic current passed down the spinal cord greatly diminished the activity of the movements, but when passed up the cord increased the amplitude and rapidity of the choreic vibrations.

Dr. Hughlings Jackson also noticed the persistence of the choreic movements after section of the spinal cord (*London Lancet*, 1875, vol. i.), but Dr. James J. Putnam (*Boston Med. and Surg. Journ.*, 1879, vol. ci. p. 690) failed to obtain the movements after section of the cord in a choreic kitten. There can be no doubt that this failure was due, as Dr. Putnam himself suggests, to the kitten being killed too soon after the section of the cord. My own experiments prove that when the division is made low down, the shock of the operation prevents the reappearance of the movements for a considerable period after the recovery of consciousness.

In the first case in which I experimented (as shown in the tracing herewith submitted), the movements were arrested by ether, and after division of the cord in the upper dorsal region did not reappear for over a quarter of an hour after complete recovery of consciousness. They first made their reappearance in the hind legs, and afterwards in the front legs. The movements were finally much more violent in the hind legs than they had been before section; but all synchronism between the front and hind legs was completely set aside by the section.

In the second experiment the movements were rhythmical, and, as is often the case, were most marked in the opposing front and hind legs. They were, as in the first case, immediately stopped by ether. The cord was cut in the lower dorsal region, and the movements reoccurred in the front leg so soon as the effect of the anæsthetic had gone off, but did not come back in the hind legs for over an hour after the return of consciousness. It is plain to my mind that the shock of section of the cord has an effect upon these movements. In the first experiment, the front legs were nearest the seat of section, and recurrence of their movements was longest put off. In the second experiment, the section was practised very low down and the hind legs were inordinately influenced. In the choreic dog, as in the child, the movements can certainly be inhibited temporarily by the cerebrum.



Tracing 4.—Showing the inhibition of movement by the galvanization of the sciatic nerve after section of the spinal cord.

This cerebral inhibition is readily explained upon the theory of Setschenow's centre in the brain, but inhibition may certainly occur independent of any cerebral action. The cord was thoroughly cut in the second animal experimented upon, as was proven not only by the paralysis of the hind legs, but also by the fact that no pain was caused by very powerful irritation of an exposed sensory nerve of the largest size. Under these circumstances, the choreic movements were very markedly inhibited by galvanization of the sciatic nerve, as is shown by the photographic reproduction of the tracing made. (See tracing No. 4.) The upper line represents the writing of the movements of the right leg, the lower line the time of application of the current. This experiment affords direct proof that there are inhibitory centres in the cord, or else that the ganglionic motor cells of the cord are directly inhibited by peripheral impulses.

From a pathological point of view, the experiments upon the section of the cord just detailed are of great importance as fixing the seat of the lesion. They have been so often repeated with such concordant results, that we must consider it settled that the movements in the choreic dog originate in the spinal cord, and that the spinal cord is the seat of the lesion; of course the observed facts do not prove that the brain ganglionic cells are in a normal condition; indeed, the actions of the choreic dog indicate that they share the abnormal condition of the spinal cells.

In 1875 (*London Lancet*, vol. i. p. 610), Dr. Gowers examined the spinal cord of a choreic dog and found no structural alteration. Two years subsequently, however, Drs. Gowers and Sankey (*Medico-Chirurgical Transactions*, London, 1877) determined the presence of lesions in the cord of another animal. They give of it the following account:

"The most conspicuous change was the infiltration of limited areas with small round 'lymphoid' cells, which varied in size, the average being .0075 mm. In most cells no nucleus was visible, but in some a large round nucleus occupied about half or two-thirds of the area of the cell. These cells were not uniformly infiltrated in the affected areas. In the white substance they aggregated in tracts, often branching and extending through part or the whole of the affected column. When the infiltration affected the gray substance it was more uniform; but here, also, the corpuscles were aggregated in masses as well as scattered. The aggregations in both gray and white matter corresponded to the course of vessels. Whenever a section divided such a tract transversely, a vessel could always be detected in its centre. Where the infiltration was extensive, the corpuscles were so densely massed that the individual cells could scarcely be distinguished; but where it was slighter, a vessel could often be seen to be encrusted with a layer of such cells. In some places a few only lay in the perivascular sheath, but in other places similar corpuscles lay outside the perivascular sheath and in the adjacent tissue. In the white substance in the neighborhood of these tracts similar cells lay among the nerve-fibres, and in some places were so numerous as to occupy a larger area than the nerve-fibres. In some places, indeed, the nerve-elements appeared to have been destroyed.

"These cells were deeply stained with carmine, and still more deeply by logwood. They resemble closely white blood-corpuscles in size and appearance. (The measurements given above correspond closely to those of white blood-corpuscles in hardened tissues.) They were identical in appearance with the white blood-corpuscles which were seen here and there among the blood-disks within the vessels. In some sections the walls of the vessels appeared to contain similar cells, as if in the process of diapedesis. From these appearances we are inclined to regard these cells as being actually white blood-corpuscles, and the morbid change as a *leucocytal infiltration.*"

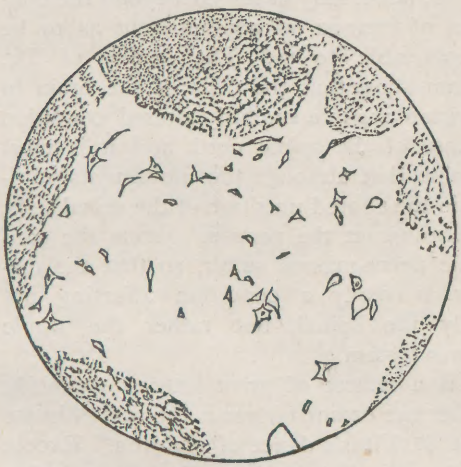
In 1879, Dr. J. J. Putnam (*Boston Med. and Surg. Journ.*, vol. ci. p. 691) examined very carefully the nervous system of two kittens which had suffered from chorea. In neither of these animals were there any alterations at all similar to those described by Gowers and Sankey. There were no abnormal leucocytes in any of the nerve-centres. In one kitten no change whatsoever could be found in any of the nerve centres. In the second there was a marked injection of the blood-vessels, and the ganglion-cells in the spinal cord, as well as those in the brain, failed to imbibe the staining fluids, and even in glycerine could not be satisfactorily studied.

I have examined the spinal cord of four dogs. Three of the dogs were killed while still able to go about. Two of them suffered from typical rhythmic chorea, one from a chorea with movements exactly simulating those of a child, and associated with great weakness. The fourth animal died of the disease, after having completely lost control

of his hind legs. In each of these cases there were numerous lymphoidal cells in the gray matter, very few, if any, of them appearing in the white matter. The cells were mostly scattered, but sometimes occurred in groups of eight or more. They were distinctly granular, and some of them were nucleated. The nuclei were usually small, but in some cases were large, nearly filling the cells. In none except dog No. 4 could any connection whatever be made out between these cells and the blood-vessels, the perivascular spaces being entirely free. In the dog that died of the disease, the cells were more numerous than in the others, and could in some instances be found in the perivascular spaces. In no instance, however, was there any such outpouring of leucocytes as is described by Gower and Sankey.

scurely arranged in three groups. Just posterior to the spinal canal, and at a little distance laterally from it, is a small group of very large cells. In the extreme anterior portion of the gray matter is a much larger group of similar cells, whilst in the lateral peripheral portions of the gray matter are numerous cells, mostly smaller than those just spoken of. Whilst this general topographical description is the most correct that can be made, I have found that the position of the cells really varies very much, not only on different levels of the cord, but in the two sides of the same section.

In my paraplegic dog the group of spinal ganglion-cells nearest the spinal canal was the most profoundly affected. As seen with a low power, the cells could not be discovered; but



Normal cord.

Choreic cord, paraplegia.

Figs. 1 and 2 are reproductions of camera drawing with low power, to show disappearance of ganglion-cells. The outline below is of edge of spinal canal.

An examination of healthy dogs shows that these lymphoidal cells are to be found in the normal cord. They are, however, not so numerous as in the severe cases of chorea examined; but I could not make out any congestion of the blood-vessels or other lesions which seemed to indicate that the outpouring of these leucocytes was the cause of the symptoms. On the other hand, my studies have clearly shown that the ganglionic cells of the cord are profoundly affected.

It has already been stated that Dr. Putnam noticed in one of his kittens that the ganglionic cells would not take the staining fluid. This represents the earliest alteration that is to be distinctly recognized. In my dog that died with paraplegia the change was more pronounced and widespread than in either of the others.

The motor cells in the dog's cord are ob-

when the higher powers of the microscope were used, it was found that their position was occupied by irregular, globose, crumpled-looking masses, without sharp outline, and taking the staining very faintly. No granulations, no nuclei, no processes were apparent. These masses certainly represent the cells in the last stages of degeneration, and were of various sizes. The cells of the other group were plainly altered. They took the staining very faintly. In a large proportion of them no nuclei could be found, and in quite a number about one-fourth of their contents was occupied by a large mass, having a sharp definite outline, free from granular contents, and having the appearance of a vacuole. Within this body was a nucleus or nucleolus, so that the body itself was probably an altered nucleus, but I have never seen anything like it in the normal cord. In many cases the pro-

cesses were gone, and the irregular borders of the cells were completely wanting in definiteness. In some instances globose masses of degenerated protoplasm, similar to those described a few lines above, occupied the places of some of the cells.

In the dog which had the choreic movements like those of St. Vitus's dance there were cells which had undergone extreme degeneration, but they were not so numerous as in the preceding case. In the sections examined it was also the peripheral cells which were most affected. It is probable that this location varies, and that more numerous sections of the two cords than I examined would show in some place in each cord one group, in other places other groups of cells were the most severely attacked.

In the milder cases of the disease, when the animal had been killed in the early stages, the only changes in the cells were the very frequent absence of the nuclei, the failure of granulations in the protoplasm, the loss of power to take staining fluids, and rarely the occurrence of sharply-defined vacuoles.

The evidence which we now have concerning the changes of the nervous system in canine chorea is sufficient to establish definite conclusions. The lesion which was found by Gowers and Sankey is evidently an accidental one, as it has not been present in one case examined by Dr. Gowers himself, and in half a dozen cases examined by other equally competent observers. This infiltration with leucocytes may be viewed as the result of a congestion of the part. Now it is well known that when functional excitement arises from congestion, the congestion precedes the functional excitement; and if the congestion were in the dog the cause of the choreic movements, it ought always to be discovered so soon as the latter were fairly established. On the other hand, functional excitement produces congestion, and in such cases the congestion follows the functional excitement, and varies very much in different cases, even when the excess of functional activity is apparently the same. If, then, the infiltration observed by Gowers and Sankey was the result of a functional excitement of the motor cells of the spinal cord, its absence in other cases is no more than ought to be expected.

It seems to me clear that the basal lesion of canine chorea is a peculiar condition of the motor cells of the spinal cord, which condition is at first, and may long continue to be, functional, but which finally may develop into organic change. It must be remembered that the distinction between functional and organic disease is a purely arbitrary one. Functional movements are the results of nutritive changes, and a functional disorder is one in which the nutritive changes have occurred, but have not sufficiently advanced to be recognized by our comparatively gross methods of study. The structure of the ganglionic nerve-cells is so complex in its ultimate nature, and yet so simple in its microscopic appearances, that it may be permanently and very seriously altered without leaving a physical trace which we can recognize. The first change in canine chorea is an altered nutrition, *i.e.*, functional excitement (or depression), of the multipolar cells of the spinal cord. This altered nutrition may continue until the structure of the cells is entirely destroyed, but it may never go beyond the condition of change so non-apparent as to be unappreciable to us. The studies of Dr. Putnam show that the cells of the brain in the kitten are in a similarly altered condition to those of the spinal cord, and it is most probable that although the movements originate in the altered nutrition of the spinal cells, yet throughout the nervous system the ganglionic nerve-masses suffer, so that the disorder is really a condition affecting not merely the spinal, but rather the whole nervous system.

It is a matter of great interest to decide whether the canine chorea is the same disease as the St. Vitus's dance of children. Except in the case of contagious diseases, it is impossible to determine with absolute positiveness that a certain disease in the animal represents a certain disease in the man, but to my mind it is plain that if canine chorea be not the same disorder as St. Vitus's dance of childhood, it is very closely allied to it.

It is true that the movements in canine chorea are usually rhythmical, whilst the movements in the child are ordinarily not so, and much has been made out of this difference. I have, however, seen dogs in which the movements were not rhythmical, but had all the *gaucherie* of the chorea of childhood, and in some cases of children the choreic movements do approximate the rhythmical type.

The points of resemblance in the two affections are very close and striking. In each disease it is the young which are especially attacked; in each the chief symptoms are those of disordered motion; each affection is connected with a constitutional disorder (rheumatism in the child and distemper in the dog); in each case the movements can be temporarily

inhibited by the will, are not accompanied with disorder of sensation, and are associated with loss of power and lowered general nerve tone; finally, in each disease arsenic is recognized as the standard remedy.

Even if canine chorea be considered essentially distinct from the St. Vitus's dance of childhood, it cannot be denied that the laws which preside over the nervous system of the dog are essentially the same as those which rule the human nervous system, and the following proposition must apply to man as well as to the dog:

Fifth.—Choreic movements may depend for their origin upon a diseased condition of the motor cells of the spinal cord, which diseased condition may not impair the structure sufficiently for the alteration to be recognized by our instruments, and hence may be functional; or may go on to the production of marked change in the ganglionic cells, or even to their total destruction, when it is entitled to rank as an organic affection.

When we approach immediately the discussion of the nature of St. Vitus's dance of childhood, we find that numerous post-mortems have been made upon this disease with very varying results. In my opinion the older autopsies ought to be disregarded. The means of investigation were so imperfect, and were so imperfectly used, that the danger of being misled by these observations is greater than the chance of receiving enlightenment. Nevertheless, it seems to me that, after throwing overboard much rubbish, there remain certain cases, in which after properly conducted autopsies no appreciable lesion could be found. The positive results which have been reached in other cases are best glanced at under two heads: First, brain alteration. Secondly, alteration of the spinal cord.

Among the most remarkable papers upon the brain-lesions of chorea is that of Dr. Broadbent, who has demonstrated that the corpus striatum and thalamus opticus are in some cases the location of the lesion. Dr. B. states that a variety of morbid conditions of these ganglia may produce chorea, but the most frequent alteration in his cases was a capillary embolism of the corpus striatum, thalmus opticus, and their vicinity. A number of autopsies have been made confirming the existence of capillary embolism in the brain in fatal chorea, and it would seem as though there was an intimate relation between chorea and this condition of the brain. The association is too frequent and too peculiar to be merely the result of a chance. On the other hand, it is perfectly evident that in many cases of chorea no such lesion exists. It is absurd to suppose that the chorea which is produced in a few hours by a fright, and is cured in a few days by arsenic, is the result of so serious an organic lesion as that indicated. Moreover, there have been an abundance of autopsies in which capillary embolism did not exist. Again, as in the case reported by Tuckwell, other changes in the brain have been noted besides those of embolism. It must, therefore, be concluded that an acute chorea may be intimately associated with minute cerebral embolism and also with other lesions of the brain; among which lesions may be especially mentioned the peculiar alteration of the ganglionic cells of the brain noted by Meynert as pervading the whole organ in a case of chorea.

In regard to the spinal cord, the following paragraph from the article of Von Ziemssen in his Encyclopædia expresses the evidence to the date of its writing, 1877: "In the spinal cord alterations have been repeatedly found, namely, hyperæmia of the medulla and the membranes, softening of the cervical and also of the dorsal medulla (Romberg, Ogle, Gray, Golgi, De Beauvais, Hine, Brown-Séquard, Lockhart-Clarke); interstitial proliferation of nuclei and hyperplasia (Rokitansky, Steiner, Meynert, Elischer), and sometimes serous exudation in the central canal, proliferation of nuclei in the adventitia of the vessels, and regressive metamorphosis in the ganglion-cells (Elischer)." Since this paragraph was penned spinal lesions have been discovered in chorea by Dr. Dickinson, by Dr. Bury, and by Dr. James Ross. According to Dr. Dickinson, the part of the cord especially affected in the disease is "the central portion of each lateral mass of gray matter comprising the root of each posterior horn."

Dr. Ross says of a case, "I was struck with the alteration presented by the accessory cells of the anterior gray horns; they appeared shrivelled, their protoplasm was granular, their nuclei were obscured, and many of their processes were indistinct or absent."

Without indulging in further quotations, it may be stated that we have the evidence of at least five or six different observers as to the alterations of the spinal ganglionic cells in acute human chorea, and that in numerous cases these alterations in all probability existed, but were not noted simply because they were overlooked. I do not mean to assert that an appreciable lesion of the spinal cells is

always to be found in St. Vitus's dance; indeed, I am quite confident that the disease may prove fatal without such lesion. Nevertheless, I am well assured that such alterations are very frequent in fatal cases, and have in not a few instances been overlooked because not appreciated and sought for.

The marked tendency of the choreic movements to affect one side of the body or one limb has led many observers to confine their careful examinations to the brain, this hemi- or monoplegic tendency being believed to show that the movements originate in the brain and not in the spinal cord. A clinical study of the various human affections of the motor ganglia of the cord shows, however, that in such disorders spinal monoplegias are not at all uncommon, and that in some cases hemiplegias may be seen. In many cases of chorea of the dog this localization of the movements in one limb or in one side of the body is quite pronounced; indeed, this was the case in dogs in which I positively determined the spinal cord to be the seat of the lesion: it cannot therefore be considered that a hemiplegic much less that a monoplegic chorea is necessarily of brain origin.

It is plain that the similarity of the lesions which have been recently noted in the ganglionic cells of the spinal cord in fatal cases of human chorea with those which I have found in the dog is very apparent, and increases the probability that the two affections are essentially the same disorder. Whether this be so or not, considerations previously detailed (see proposition five) show that choreic movements in the child, as in the dog, may originate from a diseased condition of the ganglionic spinal cell. Further, as a diseased condition of the spinal ganglionic cells has been found in several cases of human chorea, it must be allowed that such diseased state is at least one of the fundamental pathological alterations of human chorea. It is perfectly clear, on the other hand, that the disorder is not confined to these multipolar cells. The researches of Dr. Putnam, already quoted, show that in the cat alterations may be found, at least in some cases, in the brain ganglionic cells, and every clinician knows that the cerebral functions are often profoundly affected in the choreic child. The will, the intellectual and the emotional faculties are all prone to show the presence of an abnormal influence in chorea, and it seems, therefore, that we must consider that in the *choreic child the ganglionic cells in the whole cerebro-spinal system suffer, and that this alteration is the base of the disease:* or, in other words, the *morbid pathology of chorea, or St. Vitus's dance of childhood, may be said to be a diseased condition of the ganglionic structures of the cerebro-spinal axis, which abnormal state may exist without alterations of structure sufficient to be determined by the microscope, or may go on until it is accompanied with marked structural lesions.* Further, this condition must be looked upon as one of lowered tone, and it must be allowed that it may be produced by various causes, but is little prone to occur in persons of robust nervous system. The vital choreic depression of the nerve-cells may be the result of emotional disturbance, as in the chorea produced by fright. It may be the result of the influence of the rheumatic diathesis or poison upon the affected tissues. It may be the result of the partial occlusion of the circulation by cerebral capillary thrombosis or embolism, and there are probably other efficient causes in its production. The course of a chorea, as well as the severity of the symptoms, very naturally depends upon the cause of the abnormal condition, and hence we have the varieties so plainly marked in this so-called disease.